Letter to the Editor

A case description of primary pleural epithelioid angiosarcoma in an instance of sudden massive hemoptysis in a 52-year-old man

Jinyang Wen, Lianggeng Gong, Xiaoning Huang^

Department of Radiology, the Second Affiliated Hospital of Nanchang University, Nanchang, China

Correspondence to: Xiaoning Huang, MD. Department of Radiology, the Second Affiliated Hospital of Nanchang University, No. 1 Minde Road, Nanchang 330006, China. Email: haxang@163.com.

Submitted Aug 01, 2021. Accepted for publication Dec 01, 2021.
doi: 10.21037/qims-21-772
View this article at: https://dx.doi.org/10.21037/qims-21-772

Introduction

Epithelioid angiosarcoma most commonly arises in deep soft tissues, but the thyroid gland, adrenal glands, and skin have also been reported as primary sites. However, epithelioid angiosarcoma rarely occurs in the pleura. Primary angiosarcoma of the pleura is a rare and invasive tumor that accounts for about 1% of all tissue sarcomas. Herein, we report the case of a 52-year-old man with pleural epithelioid angiosarcoma who had a medical history of tuberculosis. A history of tuberculosis and pleural masses may serve as potential diagnostic clues for primary pleural angiosarcoma. By reporting this case, we hope to provide more information about pleural epithelioid angiosarcoma and improve radiologists' understanding of this disease.

Case presentation

All procedures in this study were performed in accordance with the ethical standards of the Second Affiliated Hospital of Nanchang University medical research ethics committee and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient to publish this case report and the accompanying images. A copy of the written consent form is available for review by the editorial office of this journal.

A 52-year-old man presented to the emergency department of the Second Affiliated Hospital of Nanchang University complaining of sudden massive hemoptysis that had lasted for 1 hour. The patient had a cough and had experienced expectoration for 1 month before presenting to the emergency department. Ten years earlier, he had been diagnosed with tuberculous pleurisy of the right lung, for which there is no evidence of the apparent cause, and recovered after being administered anti-tuberculosis treatment.

A chest computed tomography (CT) scan which was performed in the emergency department revealed bilateral pleural effusion, right middle and lower lobe atelectasis, and mediastinal and axillary lymphadenopathy. A huge mass measuring 17.5 cm × 11.8 cm × 11.9 cm which touched the right chest wall was also identified. The mass was an oval solid mass with uneven low density. Small areas of hemorrhage and necrosis were also observed. The lesion contained some calcification, which was located primarily on the periphery. A contrast-enhanced CT scan showed slight uneven progressive enhancement (*Figure 1*). The patient had persistent hemoptysis; urgent treatment was therefore required. According to its CT features, the bulky mass was highly likely to be a low-grade malignant tumor that derived from the patient's chest wall.

For the purposes of both diagnosis and treatment, the patient subsequently underwent a right thoracotomy and pleural decortication. Exploration during surgery found a large quantity of dark red tissue wrapped in fibrous tissue. Histopathological examination revealed the mass to be a pleura-derived epithelioid angiosarcoma. Subsequent microscopic examination showed that the tumor consisted

^ ORCID: 0000-0003-2854-3923.

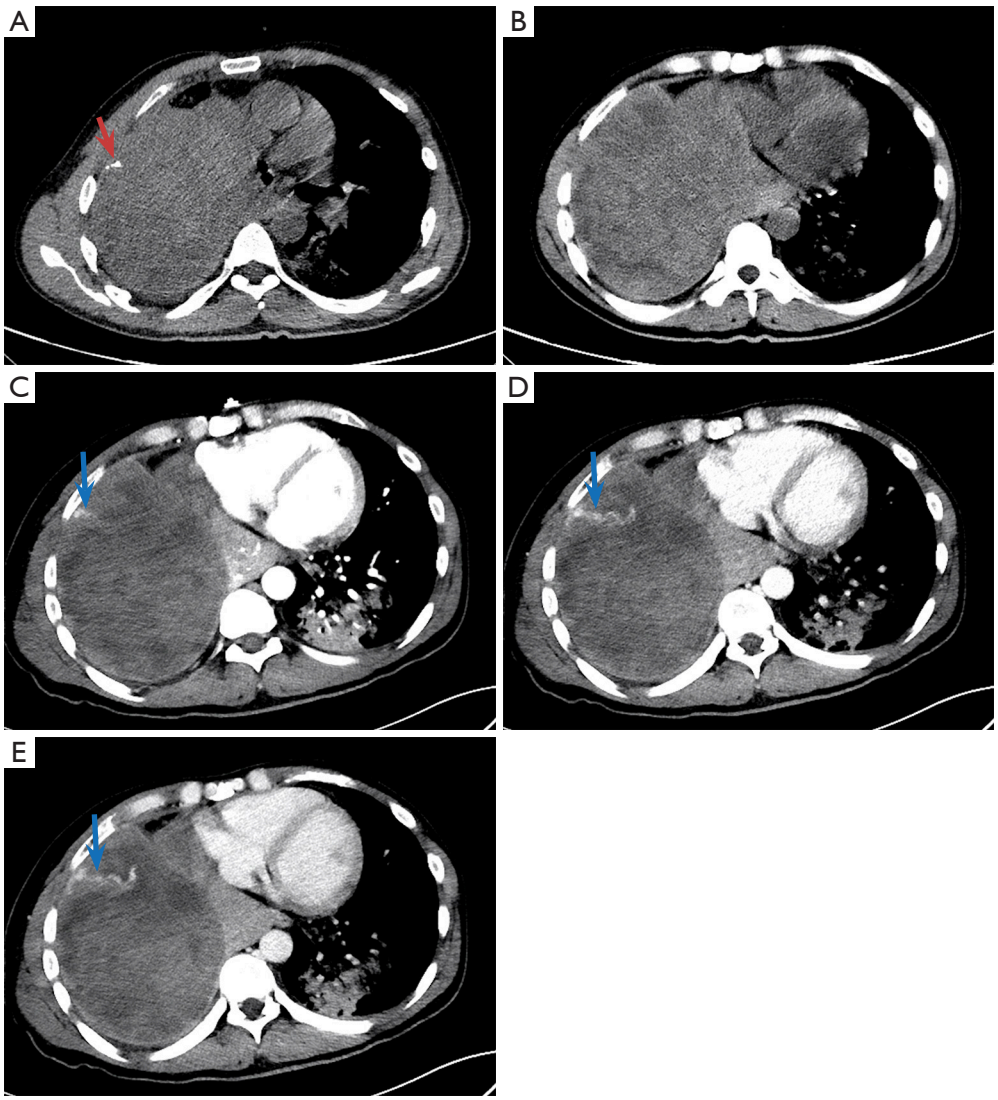

Figure 1 Axial chest computed tomography (CT) scan showing a mass in the right pleural cavity affiliated with the chest wall, with narrowing of the spacing of adjacent ribs. (A) The red arrow represents calcification located primarily on the periphery. (B) No enhanced phase was observed. (C) Arterial phase. The blue arrow represents the feeding vessel. (D) Venous phase. (E) Delay phase. Contrast-enhanced CT showed slight uneven progressive enhancement (the scanning times for the arterial phase, venous phase, and delay phase were 20 to 25 s, 40 to 50 s, and 2 min, respectively. The injection rate of the contrast media was 3.0 mL/s).

of highly atypical epithelioid cells and anastomotic blood vessels accompanied by extensive areas of hemorrhage and necrosis. Further, mesenchymal (vimentin) and endothelial (CD31 and Fli-1) markers were found to be strongly positively expressed in tumor cells. Pathological microscopic examination and the immunohistochemical features supported the diagnosis of epithelioid angiosarcoma (*Figure 2*). After surgery, the patient was treated with chemotherapy and targeted therapies (single nab-paclitaxel, methotrexate and vincristine with propranolol, anlotinib, pembrolizumab, and pembrolizumab with gimeracil). After receiving this treatment from December 2018 to October 2021, the patient is still alive currently (*Figure 3*).

Discussion

Epithelioid angiosarcoma most commonly arises in deep soft tissues. However, it rarely occurs in the pleura.

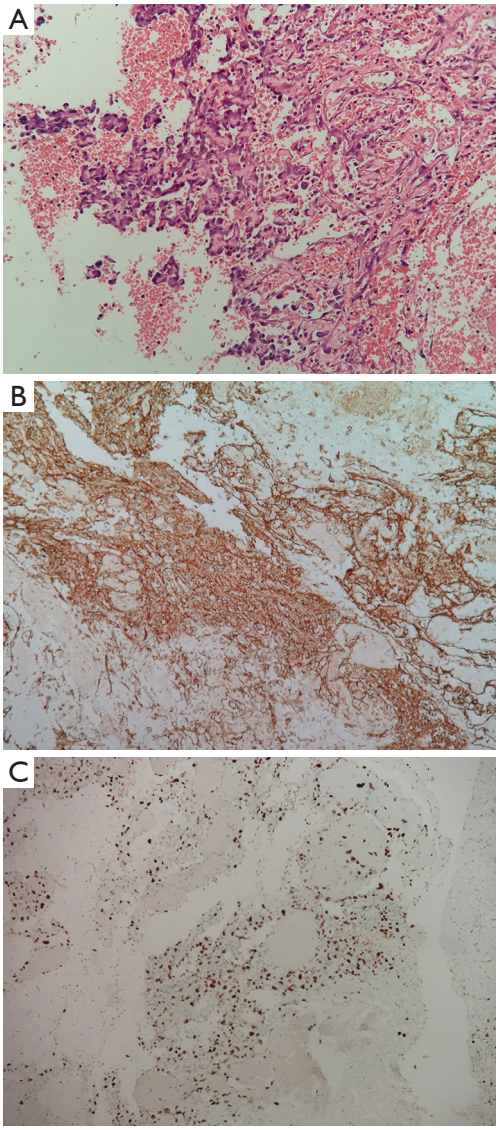

Figure 2 Neoplastic cells presenting as epithelioid in morphology (A) (hematoxylin-eosin, ×400), forming irregular anastomosing vascular channels which look like a sinusoidal network. Immunohistochemical staining (×200) results showing that the tumor cells were strongly positive for CD31 (B), and the Ki67 index was about 50% (C).

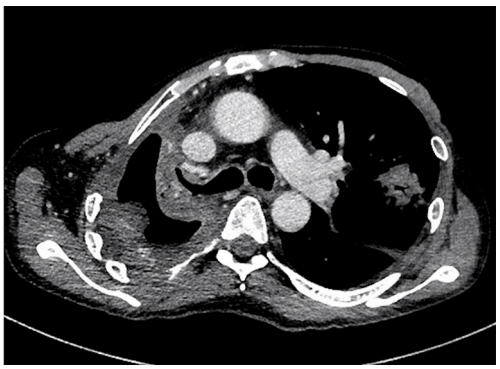

Figure 3 The latest computed tomography scan was performed on October 1, 2021, after the patient had received treatment.

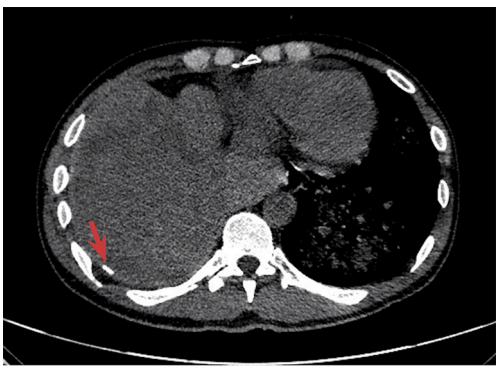

Figure 4 The computed tomography scan without contrast was performed in the emergency department. The red arrow represents pleural calcification.

Primary pleural angiosarcoma is a rare and invasive tumor that accounts for about 1% of all tissue sarcomas in worldwide (1). There are many similarities between the case described herein and those reported in the literature. First, the occurrence of pleural epithelioid angiosarcoma has previously been reported to possibly be related to chronic tuberculous empyema (2). Our patient had a medical history of tuberculosis and clinical manifestation of massive hemoptysis, and his CT examination showed calcification of pleura, which may have resulted from tuberculosis pleurisy (*Figure 4*). Moreover, the tumor exhibited uneven progressive enhancement, which was mainly located at the edge of the mass, as shown by the CT scan. This observation may be related to many of the new blood vessels present in the tumor having incomplete vascular cavities, which prevented the contrast agent from entering the center of the mass. Consequently, there was no enhancement in the center of the mass, while at the edge of the mass, there were some uninvaded blood vessels, which resulted in moderate enhancement (3).

Primary pleural epithelioid angiosarcoma should be distinguished from pleural mesotheliomas, metastatic tumor, pleura-derived neurogenic tumors, and solitary fibrous tumors (4). Pleural mesotheliomas always

present with a higher density than pleural epithelioid angiosarcomas. They are also accompanied by bone destruction and apparent strengthened pleura, and present with mild to moderate heterogeneous enhancement. A metastatic tumor of the pleura points to a history of primary malignancy. Metastatic tumors often show multiple lesions with varying sizes and shapes, which can cause rib bone destruction and pleural effusion. Neurogenic tumors of the chest wall always show as nodules or a mass with uneven density that is slightly lower than that of soft tissues, and patients are occasionally symptomless or experience only sharp pain in the chest wall. These tumors may be accompanied by cystoid degeneration and compressive bone resorption located near the ribs, and exhibit mild progressive mottled enhancement with no enhancement in the necrotic area of the cystoid. A solitary fibrous tumor of the chest wall is usually a medium or high-density mass that shows clear boundaries, a complete capsule, and prominent progressive enhancement.

In conclusion, when a mass or nodule with large areas of necrosis, hemorrhage, and uneven enhancement is found in the thoracic cavity and is accompanied by adjacent pleural thickening and lung compression, primary pleural epithelioid angiosarcoma should be considered. Nevertheless, the final diagnosis requires subsequent pathological and immunohistochemical examination.

Acknowledgments

The authors thank the patient and his family for their kind cooperation.
Funding: None.

Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-21-772/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Zhang S, Zheng Y, Liu W, Yu X. Primary epithelioid angiosarcoma of the pleura: a case report and review of literature. Int J Clin Exp Pathol 2015;8:2153-8.
2. Aozasa K, Naka N, Tomita Y, Ohsawa M, Kanno H, Uchida A, Ono K. Angiosarcoma developing from chronic pyothorax. Mod Pathol 1994;7:906-11.
3. Jiang J, Deng H, Nie J, Zhang S, Zhang M, Yu J. Imaging findings of primary epithelioid angiosarcoma of scapula: a case report. Chinese Journal of Radiology 2018,52:231-2.
4. Fan C, Liu Y, Lin X, Han Y, He A, Wang E. Epithelioid angiosarcoma at chest wall which needs to be carefully distinguished from malignant mesothelioma: report of a rare case. Int J Clin Exp Pathol 2014;7:9056-60.

Cite this article as: Wen J, Gong L, Huang X. A case description of primary pleural epithelioid angiosarcoma in an instance of sudden massive hemoptysis in a 52-year-old man. Quant Imaging Med Surg 2022;12(4):2575-2578. doi: 10.21037/qims-21-772

